# Exercise testing in children with cystic fibrosis

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*J R Soc Med* 2003;**96**(Suppl. 43):23–29 SECTION OF PAEDIATRICS & CHILD HEALTH, 26 NOVEMBER 2002

## INTRODUCTION

The medical management of paediatric patients with cystic fibrosis (CF) is multidisciplinary and most therapeutic efforts are centred on the main clinical manifestations of progressive destructive airway disease and pancreatic insufficiency. Chest physiotherapy aims to reduce airway obstruction, improve ventilation and delay the progression of the pulmonary disease process and is an integral component of care. In addition to airway clearance, advice regarding exercise, posture and maintenance of thoracic mobility[1] are also components of the physiotherapy regimen. Exercise testing, advice and prescription are no longer seen as merely complementary to airway clearance techniques but play an increasingly relevant role. Debate remains, however, as to the most effective, appropriate and useful way to determine a patient's tolerance of physical activity. This article explores some of the issues raised in this debate, discusses the practical aspects of exercise tests currently used in the paediatric CF population and offers some recommendations for clinical practice.

## THE ROLE OF EXERCISE IN THE MANAGEMENT OF CHILDREN WITH CF

Regular systemic exercise is increasingly seen as an essential part of the physiotherapy management of children with CF. It can increase cardio respiratory fitness, maintain the patient's strength, endurance and mobility and promote a good body image. Short-term studies have shown an improvement in lung function following programmes of enhanced physical activity. More recently, a randomized controlled trial of 72 children and adolescents with CF[2] reported the outcome of a 3 year home-based exercise intervention. The investigators demonstrated that the exercise programme was accompanied by a slowing of the decline in forced vital capacity (FVC) and forced expiratory volume in one second ($FEV_1$) and an improvement in the children's sense of well being.

In healthy volunteers, exercise during childhood is a major determinant of peak bone mass in adulthood.[3] With the improved survival in CF there has been greater recognition of the importance of preventative measures in childhood. These aim to allay complications such as oesteopenia and osteoporosis, which are seen in adults and adolescents with CF.[4]

## WHY TEST EXERCISE TOLERANCE IN CHILDREN WITH CF?

In health, systemic exercise is constrained by circulatory rather than ventilatory limits and by the symptoms of breathlessness or muscle fatigue. However, in children with pulmonary disease, exercise capacity is limited by the degree of severity of their lung disease.[5] With the inevitable progression of lung disease, the ability to undertake physical activity is reduced.[6] Although this reduction is associated with declining lung function, it is not always possible to predict exercise tolerance from standard pulmonary function parameters. Assessment of fitness and exercise tolerance in children with CF is a useful measure of the impact the disease is having on the patient, particularly when repeated over time. In addition, the measurement of an individual's level of exercise activity and exercise tolerance can be used to identify functional limitations and thus the effect that the disease is having on activities of daily living and quality of life. It is also useful in predicting disease prognosis,[7] it allows the safe prescription of exercise therapy, and may be a useful outcome measure in the evaluation of novel treatments.

## MEASUREMENTS OBTAINED DURING EXERCISE TOLERANCE TESTING

During exercise tolerance testing of paediatric patients with CF a number of objective and subjective measurements can be obtained for the purpose of assessment (Box 1).

### Spirometry (pre-exercise and post-exercise testing)

During tolerance testing, exercise-induced bronchospasm can be caused by bronchiolar smooth muscle contraction, stimulated by the increased ventilation associated with exercise. Higher intensity and longer duration of exercise as well as breathing cool, dry air seem to increase its occurrence. Whilst it is important to be vigilant for any symptoms which may include wheezing, shortness of breath, chest discomfort and coughing, pulmonary function

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*Box 1* **Measurements obtained during exercise tolerance testing**

| *Variable* | *Measurement tool* |
|---|---|
| Work capacity | Cycle ergometer (resistance)<br>Treadmill (speed and incline)<br>Walking (speed and distance) |
| Heart rate | Pulse meter<br>Heart monitor |
| Oxygen saturation ($SaO_2$) | Pulse oximeter |
| Pulmonary function—pre/post exercise | Spirometer |
| Respiratory rate—breaths/minute | Observation |
| On-line gas analysis | Gas analyser<br>Mass spectrometry |
| Breathlessness and muscle fatigue | Subjective—modified Borg score/visual analogue scale<br>Objective—count scores |

may be measured before and after an exercise test in order to monitor the child's bronchial responsiveness.

## Assessment of breathlessness

Breathlessness is usually defined as the subjective awareness of having difficulty in breathing or feeling out of breath. It is a normal sensation experienced on physical exertion and is related to the intensity of the exercise. It is also a common complaint of respiratory disorders and is often a distressing symptom, occurring even at rest in more severe disease.

The quantification of breathlessness is useful particularly during exercise testing and subjective tools to record/assess breathlessness are well recognized in adult practice. These include the modified Borg scale of perceived breathlessness (modified from the original Borg scale of perceived exertion[8]) and visual analogue scores (VAS) (Figure 1). In practice, scores such as the modified Borg scale are not always readily understood by children, because of the terminology used. However, both give similar scores despite their differences.[9] It is likely that many children simply use the ratings 1–10 on the Borg score and ignore the descriptors of that level, thus using the score in a similar way to the visual analogue scale.

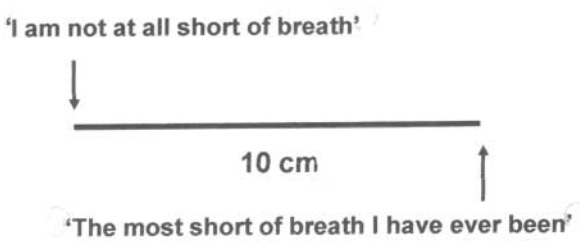


*Figure 1* **Visual analogue score (VAS)**

Two objective scores to evaluate breathlessness have also been validated. Both are counting scores, using either the maximum number of counts per second per breath[10] or counting the number of breaths taken to count to 15 in 8 seconds.[9] These are simple reproducible tests which are easy to learn and are designed to give a useful objective indication of the degree of breathlessness. However, neither takes into account the child's perception of breathlessness nor the discomfort they may be experiencing. They are therefore best used to complement one of the subjective scores rather than replace them.

## Perceived muscular effort

Exercise tolerance tests are highly effort dependant and rely on the willingness of a patient to endure the sensations of breathlessness and leg muscle fatigue. Although muscle function is thought to be normal in CF, poor nutrition and pulmonary disease can lead to decreased lower limb muscle mass. Perceived muscular effort before and after exercise testing, should be measured using both a modified Borg scale and/or by VAS, as a means of determining the reasons why exercise capacity may be diminished.

# MEASUREMENT OF EXERCISE ACTIVITY IN CHILDREN WITH CF

When making a decision on the most appropriate way to measure levels of exercise activity in an individual patient, consideration should be given to the advantages and disadvantages of each method.

## Self reported physical activity questionnaires

A number of physical activity questionnaires have been used within the paediatric CF population. Britto *et al.*[11] reported six domains of a self-administered questionnaire (the Centers for Disease Control's Youth Risk Behaviour Survey) to be a useful measure, when comparing self-reported physical activity and participation in sports activity among 116 adolescents with CF with age matched controls. Kriska's modifiable activity questionnaire[12] was found to be a useful measurement tool by Hussey *et al.*[13] when studying levels of physical activity in over 700 normal school children. Nixon *et al.*[14] also used this tool to examine the habitual physical activity of 30 children and adolescents with CF. He found them to engage in less vigorous activity than their healthy non-CF peers despite having good lung function. In addition, the use of a self-reported exercise diary in 20 patients aged 6–19 has been found to be both repeatable and to reflect global well being and general activity.[15]

### Assessment of exercise tolerance

A simple and useful way to assess functional limitation in the very young CF patient is to observe their response to routine activities of daily living such as feeding and play. Direct observation of the child's breathing pattern, colour along with measurement of oxygen saturation and heart rate, can provide valuable information relating to the child's response to physical activity.

Although this test is obviously not maximal and is dependent on patient effort and attention span, it provides useful information regarding the individual's response to exercise and has the advantage of being cheap, easy to use and it can be performed in a variety of settings including the home.

## MEASUREMENT OF EXERCISE TOLERANCE IN CHILDREN WITH CF

Clinical exercise tolerance tests fall into two main categories, that of the more formal laboratory-based exercise tests and the field based clinical exercise tests.

### Laboratory tests

The most precise method of assessing exercise capacity in any subject group is by formal laboratory testing with online analysis of expired air. This method will provide the most commonly used physiological measure of a subject's exercise capacity, which is oxygen consumption ($VO_2$).

$VO_2$ readings directly reflect the supply of oxygen by the cardiorespiratory system and its uptake by working muscles. The highest recorded $VO_2$ value is generally referred to as the $VO_2$ max. This is the measurement made following a maximal exercise test, and represents the efficiency of the cardiorespiratory system to deliver $O_2$ to working muscles and the ability of those muscles to endure sustained use.[16] However, identification of true $VO_2$ max depends on the measurement obtained fulfilling certain criteria. If these criteria cannot be achieved, then maximum oxygen uptake should be designated as $VO_2$ peak.

Progressive maximal tests using a cycle ergometer or treadmill provoke a maximal response in patients and provide data relating to the individual's physiological response to an increasing workload and their exercise capacity.

Measurements undertaken include:

- Spirometry (pre-test and post-test)
- $VO_2$
- $VCO_2$
- Maximum ventilation
- Oxygen saturation throughout the test
- Heart rate and ECG recording
- Subjective/objective measures of breathlessness
- Muscle fatigue.

The disadvantages of these tests are that they are not really representative of patterns of day-to-day physical activity particularly as experienced by children. Laboratory testing is expensive in terms of time, personnel and equipment and may not offer any advantage over some of the simpler field tests in terms of day-to-day clinical management. In addition, they can be extremely stressful and many patients with CF are reluctant to perform them on a routine basis.

### Field tests

There is debate as to which is the most appropriate protocol for informal or field-based exercise testing. While field-based exercise tests do not always determine a subject's maximal exercise response, they can give valuable clinical information on functional limitation.

With a number of tools to measure exercise tolerance available to the CF clinician, the most appropriate choice of test for an individual patient will depend on a number of considerations. These will include the type of information or data that is required and the facilities for exercise testing available to the clinician, as most units do not have access to exercise laboratories. Time, staffing and cost may also be important factors. Similarly, a patient's age and disease state may influence the decision on the most appropriate test.

### Walking tests

The 12 minute walking test was one of the first un-paced tests used to assess functional exercise tolerance in adults with respiratory disease[17]. A reduction in the time of the test to 6 minutes was later found to have no disadvantage[18] and these tests are now commonly used in the assessment of functional exercise tolerance.[19]

Walking tests are very simple to perform with subjects being asked to cover as much ground as possible in the time permitted over a marked course (usually a corridor). Measurements undertaken include:

- Spirometry (pre- and pos-test)
- Baseline and highest pulse rate
- Baseline and lowest oxygen saturation
- Subjective/objective measures of breathlessness
- Distance walked
- Muscle fatigue

The distance walked during a 6 minute walk test has been reported to correlate with $VO_2$ peak in children with severe respiratory disease undergoing assessment for heart–lung transplantation.[20] In children with mild to moderate CF, Gulmans *et al*.[21] also reported a significant correlation between the walking distance and $VO_2$ max. This study also reported good reproducibility in their group of children, although the number of subjects was relatively small. Distance covered during a walk test is commonly used in adults as a prognostic indicator for assessment of suitability for heart–lung transplantation.[22] In children, however, this may be a less useful tool for this purpose[23] possibly because the test is self-paced and the results can vary due to the influence of motivation and encouragement.

Such tests are cheap, simple, require little in the way of equipment and can be used in an ambulatory setting (Figure 3). However, they do require adequate (and uninterrupted) space and are effort dependent. This does mean that the child's attitude and motivation are very influential in determining the distance walked.

## Step tests

The 3-minute step test was adapted from the original Master two-step exercise test[24] and has been validated for use in children over 6 years of age as a means of assessing sub-maximal exercise tolerance.[25] Subjects step up and down on a standard, commercially available aerobic step (15 cm) for 3 minutes. The stepping height and frequency are kept constant and the test is paced at 30 steps/minute by the use of a metronome (set at 120). A stopwatch is used to time the test and standardized encouragement and an indication of how far the test has progressed given at each minute interval. Subjects are taught how to change the lead leg while stepping to avoid unilateral muscle fatigue. The use of a cushioned mat on the floor is useful to reduce the impact particularly to the knees.

Measurements undertaken include:

- Spirometry (pre- and post-test)
- Baseline and highest pulse rate
- Baseline and lowest $SaO_2$
- Subjective and objective measurement of breathlessness (visual analogue scale and 15 count score) before and immediately after the test
- Number of steps taken if test not completed and reason for stopping
- Muscle fatigue.

When compared with the 6-minute walk this test was shown to produce significantly greater changes in heart rate and breathlessness along with a comparable fall in $SaO_2$.[25] In children with severe lung disease undergoing assessment for heart–lung transplantation the 3-minute step test produced a significantly greater fall in $SaO_2$ than the 6 minute walk.[26] If this test is able to discriminate between those with normal resting $PaO_2$ and good exercise tolerance, and children with lower resting $PaO_2$ and poor exercise tolerance, it may prove a useful tool in the assessment of children for heart–lung transplantation as a prognostic indicator.


*Figure 2* **A patient undertaking a walk test.** During the test the operator monitoring the patient's response walks behind the subject in order to avoid influencing the patient's walking speed

The height and rate are kept constant in this test and therefore the workload undertaken does vary between subjects (as the work is a product of step height and rate as well as the subject's weight and height). In order to standardize the workload it would be necessary to adjust the step height according to subject's height and weight. In practical terms this would make the test rather more cumbersome and may not be important in the context of the way the test is used. Interpretation of longitudinal results in an individual should, however, bear this in mind particularly if they span a pubertal growth period. The test has been used to assess the effects of intravenous antibiotics on exercise tolerance in children with CF and reported to be sensitive to changes after such a therapeutic intervention.[27] However, most children with CF, particularly those

with mild and even moderate disease are able to complete the test without difficulty. It is not a test designed to measure maximal functional capacity and in milder disease the test is often completed without any detrimental effect on oxygen saturation. Therefore the nature of this test does not necessarily lend itself to be used as an outcome measure in interventional studies. However, the 3-minute step test is a useful measure of exercise tolerance in terms of heart rate response, $SaO_2$ and breathlessness during and after an exercise challenge.[28]

The advantages of this test are that it is quick to perform, easy to learn, portable and requires little space. More importantly, it is externally paced and therefore not influenced by motivational factors—as with all exercise tests, the patient can stop the test at any time should they wish.

Rebovich *et al.*[29] have described a maximal progressive step test in which workload is initially increased by a rise in frequency of stepping and then by progressive loading of weights to a vest worn by the subject. While such a test could be an extremely useful tool, the physical and physiological effects of loading the thorax during physical exercise need to be explored before this test can be widely accepted.

### Shuttle tests

The shuttle walking test (SWT)[30] is an incremental, externally paced, informal exercise test, which overcomes many problems associated with other field-based exercise tests. It can be performed as either the original 12-level shuttle walk or as a modified 15-level shuttle test. Both have been studied in adult and paediatric CF populations, but it is the modified shuttle walking test (MSWT) that has been shown to be a valid measure of exercise tolerance, as well as a reliable and sensitive measure of exercise capacity in adults with CF by Bradley and colleagues.[31,32] However the recognition that a number of physiological differences exist between adults and children[33] means it is not always possible to extrapolate findings from adults to paediatrics. The validity of the MSWT has been studied in 14 children aged 10–19 years and a high correlation between the subjects' $VO_2$ peak and distance achieved during the MSWT has been found. This suggests that it is also a valid test of exercise tolerance in paediatric CF patients.[34]

Measurements undertaken include:

- Spirometry (pre and post-test)
- Distance achieved in metres
- Heart rate
- Oxygen saturation
- Subjective/objective measures of breathlessness
- Muscle fatigue

Although the MSWT has been shown to provoke a symptom limited response in paediatrics, its disadvantages are that it remains effort dependant, requires at least 15 m of uninterrupted space to perform and, whilst it can be used as a field test, is more commonly used within the hospital setting.

Figure 3 gives a diagrammatic representation of the shuttle walk course where the patient is required to walk or run at increasing speeds back and forth along a 10 m course. The pace is set by a prerecorded audio signal which indicates when the patient has to reach each end of the course. Patients continue with the test until they are unable to carry on or if they fail to maintain the set pace.

### Cycle ergometry and treadmill tests

Progressive maximal tests using a cycle ergometer or treadmill can also be used in a clinical setting (other than an exercise laboratory) and provoke a symptom-limited response to exercise when used as a field based test.[35] There are a number of protocols that can be used for testing but the one most often quoted is the Godfrey protocol[36] which uses 10, 15 and 20 w increments per minute for children $<125$ cm, 125–150 cm and $>150$ cm, respectively.

Measurements undertaken include:

- Spirometry (pre- and post-test)
- Work load
- Peak heart rate
- Oxygen saturation
- Subjective/objective measures of breathlessness
- Muscle fatigue

## RECOMMENDATIONS FOR CLINICAL PRACTICE

- It is important to recognize that for children the ability to take part in exercise activity is a fundamental aspect of human existence[37] and that the loss of this ability has a detrimental effect on an individual's quality of life
- All paediatric patients with CF should have some form of exercise tolerance testing done at their annual

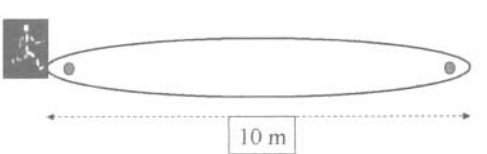


*Figure 3* **A diagrammatic representation of the shuttle walk course.** Subject walks/runs between markers. The pace is set by a pre-recorded audiotape and speed is increased at each level. The test is stopped when the patient is unable to keep the set pace or is no longer able to continue

assessment as stated in the guidelines for the physiotherapy management of CF produced by the Association of Chartered Physiotherapists in CF (ACPCF)[38]

- Prior to any exercise advice or prescription being given to a paediatric patient an exercise tolerance test should be performed as part of an exercise risk assessment procedure
- The choice and application of any exercise tolerance test should be individualized to the requirements of each child with CF and the resources of the CF unit.

## ADHERENCE TO REGULAR EXERCISE

The benefits of regular exercise are not immediate but are both noticeable and measurable over a relatively short period of time.[39,40] However, any beneficial effects are lost within a short time of regular activity being discontinued.[41] Adherence to exercise regimens is therefore a key issue. Several factors influence adherence to physical therapy in CF including age, clinical status as well as factors associated with behaviour (e.g. motivation) and those external to the patient (access and availability).

In order to improve and maintain adherence to any regular exercise programme it is necessary that the patient and family are made aware of its positive effects both in the short and long term. The child needs the support of both the family and the healthcare team. They should be given a lot of encouragement and a choice of activities including some that can be undertaken and enjoyed with family and friends.

## CONCLUSION

Exercise regimens should be adapted as health status changes[42] and the measurement of a child's tolerance of physical activity by means of exercise tolerance testing should provide the foundation for any exercise advice and prescription by the physiotherapist. It is also hoped that regular exercise testing will help emphasize the importance of activity to children with CF and their families.

## REFERENCES


1 Peebles A. Physiotherapy. In: Hill CM, ed. *Practical Guidelines for Cystic Fibrosis Care*. London: Churchill Livingstone, 1998:41–63

2 Schneiderman-Walker J, Pollock SL, Corey M, *et al*. A randomised controlled trial of a 3-year exercise program in cystic fibrosis. *Journal of Pediatrics* 2000;**136**:304–10

3 Boot AM, de Ridder MA, Pois HAP. Bone mineral density in children and adolescents: relation to puberty, calcium intake and physical activity. *J Clin Endocrinol Metab* 1997;**82**:57–62

4 Conway SP, Morton AM, Oldroyd B, *et al*. Osteoporosis and oesteopenia in adults and adolescents with cystic fibrosis: prevalence and associated factors. *Thorax* 2000;**55**:798–804

5 Webb AK, Dodd ME, Moorcroft J. Exercise and cystic fibrosis. *J R Soc Med* 1995;**88**(suppl 25):30–6

6 Nixon P, Orenstein D. Exercise testing in children. *Pediatr Pulmonol* 1988;**5**:107–22

7 Nixon PA, Orenstein DM, Kelsey SF, Doershuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. *N Engl J Med* 1992;**327**:1785–8

8 Borg G. Psychophysical bases of perceived exertion. *Med Sci Sports Exercise* 1982;**14**:377–81

9 Prasad SA, Randall SD, Balfour-Lynn IM. Fifteen-count breathlessness score: an objective measure for children. *Pediatr Pulmonol* 2000;**30**:56–62

10 Orenstein DM, Holt LS, Rebovich P, Campbell T, Nixon P. Measuring ease of breathing in young patients with cystic fibrosis. *Pediatr Pulmonol* 2002;**34**:473–7

11 Britto MT, Garrett JM, Konrad TR, Majure JM, Leigh MW. Comparison of physical activity in adolescents with cystic fibrosis versus age matched controls. *Pediatr Pulmonol* 2000;**30**:86–91

12 Kriska AM, Knowler WC, Laporte RE, *et al*. Development of a questionnaire to examine relationships of physical activity and diabetes in Pima Indians. *Diabetes Care* 1990;**13**:401–11

13 Hussey J, Gormley J, Bell C. Physical activity in Dublin children aged 7–9 years. *Br J Sports Med* 2001;**35**:268–72

14 Nixon PA, Orenstein DM, Kelsey SF. Habitual physical activity in children and adolescents with cystic fibrosis. *Medic Sci Sports Exercise* 2001;**33**:30–5

15 Rogers D, Doull IJM. Repeatability of a self-reported exercise questionnaire in stable paediatric patients with cystic fibrosis. *Netherlands J Med* 1999;**54**:S128

16 Morgan MDL. Exercise testing. In: Hughes JMB, Pride NB, eds *Lung Function Testing—Physiological Principles and Applications*. London: WB Saunders 1999:133–51

17 McGavin CR, Gupta SP, McHardy GJR. Twelve-minute walking test for assessing disability in chronic bronchitis. *BMJ* 1976;**i**: 822–3

18 Butland RJA, Pang J, Gross ER, *et al*. Two-, six-, and 12-minute walking tests in respiratory disease. *BMJ* 1982;**284**:1607–8

19 Noonan V, Dean E. Sub-maximal exercise testing: clinical application and interpretation. *Physical Ther* 2000;**80**:782–807

20 Nixon PA. Role of exercise in the evaluation and management of pulmonary disease in children and youth. *Med Sci Sports Exercise* 1996;**28**:414–20

21 Gulmans VAM, van Veldhoven NHMJ, de Meer K, Helders PJM. The six-minute walking test in children with cystic fibrosis: reliability and validity. *Pediatr Pulmonol* 1996;**22**:85–9

22 Kadikar A, Maurer J, Keston S. The six-minute walk test: a guide to assessment for lung transplantation. *J Heart Lung Transpl* 1997;**16**:313–19

23 Aurora P, Wade A, Whitmore P, Whitehead B. A model for predicting life expectancy of children with cystic fibrosis. *Eur Respir J* 2000;**16**:1056–60

24 Master AM, Oppenheimer ET. A simple exercise tolerance test for circulatory efficiency with standard tables for normal individuals. *Am J Med Sci* 1929;**177**:233–43

25 Balfour-Lynn IM, Prasad SA, Laverty A, Whitehead BF, Dinwiddie R. A step in the right direction: assessing exercise tolerance in cystic fibrosis. *Pediatr Pulmonol* 1998;**25**:278–84

26 Aurora P, Prasad SA, Balfour-Lynn IM, Slade G, Whitehead B, Dinwiddie R. Exercise tolerance in children with cystic fibrosis undergoing heart lung transplantation assessment. *Eur Respir J* 2001;**16**:293–7

27 Pike SE, Prasad SA, Balfour-Lynn IM. Effect of intravenous antibiotics on exercise tolerance (3-minute step test) in cystic fibrosis. *Pediatr Pulmonol* 2001;**32**:38–43

28 Orenstein DM. Exercise testing in cystic fibrosis. *Pediatr Pulmonol* 1998;**25**:223–5

**29** Rebovich PJ, Orenstein DM, Holt LS, Nixon PA. A comparison of aerobic fitness determined by stair steps and cycle ergometry in patients with cystic fibrosis (CF) [Abstract]. *Pediatr Pulmonol* 2000;(suppl 20);**30**:3

**30** Singh S, Morgan M, Scott S, *et al*. Development of a shuttle walking test of disability in patients with chronic airways obstruction. *Thorax* 1992;**47**:1019–24

**31** Bradley J, Howard J, Wallace E, Elborn S. Validity of a modified shuttle test in adult cystic fibrosis. *Thorax* 1999;**54**:437–9

**32** Bradley J, Howard J, Wallace E, Elborn S. Reliability, repeatability and sensitivity of the modified shuttle test in adult cystic fibrosis. *Chest* 2000;**117**:1666–71

**33** Cooper D. Rethinking exercise testing in adults with cystic fibrosis. *Am J Respir Crit Care Med* 1995;**152**:1154–7

**34** Rogers D, Smith P, John N, Oliver W, Doull IJM. Validity of a modified shuttle walk test as a measure of exercise tolerance in paediatric CF patients. *J Cystic Fibrosis* 2002;**1**(suppl 11):139

**35** Bird SR, Smith A, James K. Physiological and psychological assessments. In: *Exercise Benefits and Prescription*. Cheltenham: Stanley Thomas, 1998:107–43

**36** Godfrey S, Mearns M. Pulmonary function and response to exercise in cystic fibrosis. *Arch Dis Child* 1971;**46**:144–51

**37** Cooper CB, Storer TW. Purpose. In: *Exercise Testing and Interpretation: a Practical Approach*. Cambridge: Cambridge University Press, 2001: 1–14

**38** Association of Chartered Physiotherapists in Cystic Fibrosis. *Standards of Care For the Physiotherapy Management of Cystic Fibrosis*. Bromley: Cystic Fibrosis Trust, 2002

**39** Cerny FJ. Relative effects of bronchial drainage and exercise for in-hospital care of patients with cystic fibrosis. *Physical Ther* 1989;**69**:633–9

**40** Sevadurai H, Van Asperen PP, Cooper PJ, Mellis CM, Blimpkie CJ. A randomized control study of in-hospital exercise training programmes in children with cystic fibrosis [Abstract]. *Pediatr Pulmonol* 1999;**19**(suppl):433

**41** Zach M, Oberwaldner B, Hausler F. Cystic fibrosis: physical exercise versus chest physiotherapy. *Arch Dis Child* 1982;**57**:587–9

**42** Prasad SA, Cerny FJ. Factors that influence adherence to exercise and their effectiveness: application to cystic fibrosis. *Pediatr Pulmonol* 2002;**34**:66–72